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PART III

Operative Therapy for Ulcerative Colitis and Crohn's Disease

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IT IS NOW GENERALLY ACCEPTED that ulcerative colitis is a disease primarily and solely of the large intestine whereas Crohn's disease may affect the alimentary tract from the esophagus to the rectum. Crohn's disease of the colon (granulomatous colitis) is common either in association with small bowel disease or affecting the large bowel alone. In our experience the extent of small bowel involvement is more limited in extent than that found in regional enteritis without colonic involvement. The pathogenesis, clinical features and radiological findings of these two disease entities have already been discussed. The implications of surgical operation for these two diseases is simple for ulcerative colitis but complex in the case of Crohn's disease. Since ulcerative colitis is confined to the colon, it can be eliminated when indicated by eradication of the large intestine. Not so for Crohn's disease, since it can affect any portion of the alimentary tract and recurrence is commonplace after resection. However, it is of interest that the surgical management of Crohn's disease of the colon fares better than the surgical treatment of regional enteritis. This will become evident as we go into more detail regarding the surgical management of Crohn's disease.

Indications for Operation

The indications for operative intervention for these two diseases are similar in some respects. When either disease becomes refractory to medical therapy that has included salicylazosulfapyridine (Azulfidine®), systemic or local use of corticosteroids or immunosuppressive therapy, operative intervention is indicated. Similarly, significant retardation of growth and maturation in adolescence either due to the disease process or to the drug therapy is a most important criterion for surgical treatment. Likewise, complications from prolonged administration of corticosteroids or immunosuppressive agents in adults should direct these patients to the surgeon.

When we examine the complications of the two disease processes, we find differences that have important implications with regard to surgical management. In ulcerative colitis we may encounter uncontrollable hemorrhage, toxic megacolon, perforation, progressive liver disease, and the risk of carcinoma with long standing disease—all indications for operation. On the other hand, bowel obstruction and internal fistulization are uncommon. The complications of Crohn's disease that require operation include intestinal obstruction due to cicatricial stenosis, and enterovesical, entero-vaginal and entero-cutaneous fistulae. Fistulae between loops of small bowel or small bowel and colon do not usually indicate surgical intervention. Massive bleeding is uncommon, as is free perforation and the development of carcinoma.

In addition to the specific indications discussed above, I hasten to add that it is important to assess the patient as a whole individual, not merely to examine his disease process. It is important to know whether he (or she) is capable of carrying on his normal work and activities, can enjoy a reasonably palatable diet, is not spending his savings on expensive medications for long periods of time, and is able to enjoy life in our customary fashion.

Preoperative Preparation for Elective Resection of the Colon

In addition to the routine history, physical examination and laboratory studies, all patients should have an electrocardiogram, chest film and liver function tests including determination of prothrombin time. Serum albumin deficits should be corrected with daily intravenous human albumin, and prothrombin deficiencies with vitamin K. Anemia, if present, requires correction with whole blood or packed cells before operation is done.

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Cystoscopy may be required in cases of bladder involvement by fistula. Since many patients are receiving or have been receiving corticosteroids, adequate steroid therapy must be given before, during and following operation.

The local preparation of the colon is an important aspect of preoperative management since it is highly desirable to have a clean, empty colon as free of pathogenic organisms as possible. Most surgeons are in agreement that mechanical preparation is as important or more important than the sterilization by antibiotics. The accomplishment of mechanical cleansing requires the use of a clear liquid diet for three to four days before operation. More recently, the use of elemental amino acid diets has been greeted with enthusiasm in bowel preparation. Adequate purging of the bowel with suitable laxatives is essential, as are cleansing enemas. I favor the use of neomycin or kanamycin for 48 to 72 hours before operation to reduce the bacterial flora of the colon. It should be remembered that most anaerobic organisms are still present in significant numbers after such antibiotic preparation.

Before operation a Foley catheter is inserted routinely to monitor the urinary output and to protect the bladder during the surgical procedure. A nasogastric tube is also inserted to decompress the stomach during operation and after operation.

Preoperative Preparation for Emergency Resection of the Colon

Partial or total colectomy in emergency may be necessitated by acute obstruction, massive hemorrhage, or by perforation or the development of toxic megacolon unresponsive to conservative therapy. Under these conditions it becomes impossible to carry out a careful preparation of the colon. It is imperative to correct any volume deficits with blood or albumin or both and to initiate vigorous antibiotic therapy. Central venous pressure monitoring is essential in these critically ill patients.

Operative Management of Ulcerative Colitis

Since removal of the entire colon is curative for ulcerative colitis, total procto-colectomy is the procedure of choice. A minimal length of ileum is removed, with the proximal end brought out through a pre-determined site in the right lower quadrant as a permanent ileostomy. The ileum is everted on itself and the edge sutured to the skin so that it projects between 1 and 2 cm above the skin level. It is important to suture the ileal mesentery to the lateral parietal peritoneum to prevent internal herniation or prolapse.

More recently, there is increasing interest in Kock's[1] operation in which a large pouch is made of the distal ileum. The pouch hangs in the pelvis and exits by a very small ileostomy. This pouch is capable of holding 12 to 24 hours' accumulation of ileal contents and is catheterized every 12 to 24 hours. Removal of the rectum is accomplished either via a combined abdominoperineal approach or through the abdominal incision alone if the pelvic inlet is adequate. In males it is essential that dissection stay as close to the rectum as possible to avoid impotence. Surgical mortality ranges from 3 to 5 percent for elective operations.

Although some surgeons advocate ileorectal anastomosis when the rectum appears uninvolved with disease, the results in this country with this procedure have not been satisfactory.[2] It is my opinion that in many such cases the diagnosis is incorrect and that the patients have Crohn's disease rather than ulcerative colitis. The high incidence of recurrent disease, the risk of the development of carcinoma, and the tendency to frequency of bowel movements argue against this procedure except in rare cases.

More recently, sphincter-saving procedures have been devised and utilized successfully. Preservation of the distal anorectal segment from which the mucosa has been removed serves as a muscular tube which the distal ileum is pulled through and anastomosed to the cutaneous cuff of the anorectal line.[3] Further experience with procedures such as this is needed to evaluate the long term results.

Since one occasionally notes sparing of the right colon at the time of operation, it is tempting to preserve the right colon and to perform an end colostomy, thus avoiding ileostomy. In our experience the preserved right colon almost always becomes involved with disease, requiring right colectomy and ileostomy later. The pronounced reduction in ileostomy complications that has been brought about by the improvement in the technical aspects of constructing permanent ileostomies and the advances in ileostomy care contraindicate preserving the proximal colon in this disease.

When emergency operation is required for complications as severe hemorrhage, perforation or toxic megacolon, operative procedures less than total procto-colectomy may be indicated. It may be prudent to leave the rectum for subsequent removal rather than to prolong the operation unduly. Occasionally, diverting ileostomy has proved lifesaving in patients with toxic megacolon in whom a more extensive operation would not be tolerated. Turnbull[4] has advocated definitive ileostomy and transverse colostomy to decompress the dilated colon when walled-off colonic perforations are discovered at laparotomy. Selection of the appropriate procedure may reduce a recognized mortality of 30 to 40 percent to acceptable levels.

Operative Management of Crohn's Disease

Regional enteritis. In contrast to chronic ulcerative colitis that is cured by surgical eradication, Crohn's disease is characterized by its tendency to recur in a significant number of patients after operative therapy. Despite a recurrence rate of roughly 50 percent after operation for regional enteritis, we must not fail to recognize that an equal percentage of patients are cured by operative intervention. Furthermore, when the disease affects the colon primarily, the results are significantly better than for operations for small bowel disease.

Regional enteritis may present acutely as acute appendicitis; and when the patients are found to have a diseased ileum, resection is usually not necessary, since acute ileitis generally denotes the disease is in early phase. The appendix can be removed safely, provided the cecum is not also involved by the inflammatory process. In the latter circumstance, the risk of fistula formation is great if the appendix is removed. It is also important when terminal ileitis is present to culture material from the appendix for the presence of Yersinia enterocolitica or Yersinia pseudotuberculosis and to do the appropriate serological test. It is usually not possible to differentiate between these two forms of terminal ileitis. It is also of interest that signs of Crohn's disease have not developed in patients with Yersinia infection who were followed for periods up to five years.[5]

The surgical management of those complications of regional enteritis discussed earlier has usually consisted of resection or by-pass by exclusion. Simple by-pass by ileo-colostomy (side-to-side) has been abandoned since it does not result in complete diversion of the fecal stream and may result in the formation of a blind loop. Most surgeons favor resection as the procedure of choice when it can be safely carried out. The limits of resection tend to vary according to the preferences of the surgeon. It has been shown that radical excisions that remove more than 6 inches of normal-appearing ileum proximal to the grossly involved bowel have not lowered the recurrence rates as compared with more conservative resections. In addition, it has often been customary to remove the ascending colon, when uninvolved, in order to accomplish an ileo-colostomy anastomosis easily. I believe it is unnecessary to resect more than 6 inches of grossly normal-appearing ileum and the entire right colon. An ileo-cecostomy or end-to-end ileo-ascending colostomy are preferred, since it may be important to preserve as much of the right colon as possible to avoid postoperative diarrhea. If fistulae exist between loops of bowel, enterovesical or enterocutaneous, resection of the involved segment of bowel along with the fistulous tract is indicated. When the bladder is involved, the wall should be repaired with sutures placed through healthy tissue, and an indwelling catheter inserted for drainage.

Exclusion by-pass procedures are reserved for the occasional cases in which there is tremendous thickening and shortening of the mesentery, making resection dangerous. We have recently modified the operation where there has been extensive disease involving the entire ileum with or without skip lesions more proximal, particularly in adolescent patients. The segment of intestine bypassed has its proximal end closed and its distal end at the ileo-cecal junction transected and brought out the right lower quadrant as a mucous fistula. This modification insures a complete bypass. That is, it prevents any reflux of colonic materials into the diseased ileal segment. The rationale for this procedure is based on the striking remission that follows diversion by ileostomy for Crohn's disease of the colon.

When "skip areas" of disease are present, they may be resected along with the terminal ileitis if in reasonable proximity, but when they occur in the jejunum it is usually wise to leave them and resect only the severely diseased portion of the ileum. A conservative approach is usually justified, since one wishes to avoid the development of

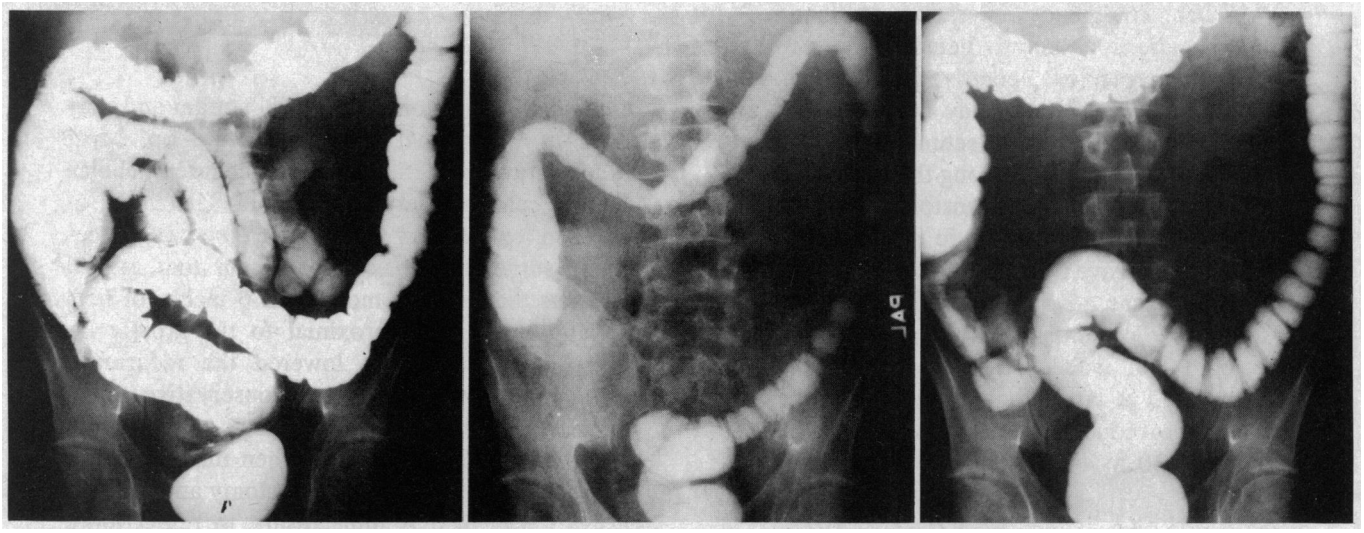

Figure 1.—Left, discontinuous and asymmetrical irregularity of the contour of the colon, most prominent in the proximal transverse and descending colon. **Center,** evidence of healing with absence of ulceration two years following diversion. **Right,** colon normal four years after ileoileal reconstruction.

a short bowel syndrome, with diarrhea and inadequate absorption.

When Crohn's disease involves the duodenum, it may be necessary to perform a gastrojejunostomy for duodenal obstruction. We have seen Crohn's disease of the duodenum in association with duodenal ulcer disease and have achieved excellent results with vagotomy and pyloroplasty. Gastric involvement may require resection if the distal stomach becomes obstructed by granulomatous disease.

Crohn's disease of the colon. Crohn's disease of the colon may be diffuse in its involvement, or it may be segmental or, rarely, limited to the rectum. In addition, any of the forms above may be associated with involvement of the terminal ileum. In my experience, roughly 40 percent of patients with Crohn's of the colon also have ileal disease. It is of some interest that, in contrast to regional enteritis without colonic disease, ileal involvement has often been of limited extent.

Where the disease appears segmental, often with stricture formation, local excision with end-to-end anastomosis will yield satisfactory results in only 50 percent of patients, the disease recurring later in the other 50 percent. However, the results appear better in patients with right sided disease and with a normal left colon and rectum. Many surgeons will elect colectomy with ileo-transverse colostomy as the procedure of choice in these patients. The accepted therapy for diffuse disease of the colon is total procto-colectomy, with ileostomy proximal to ileal disease if present. Recurrent disease at the stoma, with retrograde extension, occurs in about 15 to 25 percent of patients. Since the rectum may be spared in Crohn's disease, it is often tempting when colectomy is indicated to do an ileo-rectal anastomosis, avoiding ileostomy. However, the long term results indicate a roughly 50 percent rate of recurrent disease—similar to the results of segmental colonic resections and those done for regional enteritis.

Despite the excellent results of total procto-colectomy for diffuse Crohn's disease of the colon, for the past decade we have been interested in the effects of diverting ileostomy. Our reports in 1968 and 1971[6] emphasized the immediate and dramatic clinical response seen in all patients following diversion. Objective improvement in the radiological appearance of the colon toward normal was also observed (Figure 1). It seemed most reasonable to recommend this approach in young patients rather than to remove the entire colon. Fifty percent of our reported cases were in patients under the age of 20, most of whom had the nutritional complications of the disease or its treatment with corticosteroids. Although we achieved excellent response in all 24 patients initially, eight (33 percent) of the group had recurrence of symptoms anywhere from six months to three years after the diversion operation and total procto-colectomy was necessary (Table 1). Nine patients (38 percent) remain asymptomatic for periods of from one to six years—unwilling to risk re-connection. The remaining seven patients (29 percent) have had intestinal continuity re-established (Table 2). Three patients of the group remain well (up to eight years after operation).

TABLE 1.—*Results of Ileostomy Diversion (24 Patients)*

24	Excellent initial response
16	Continued remission (12 to 72 months)
7	Re-united (24 to 36 months after by-pass)
9	Remain asymptomatic (12 to 72 months)
8	Relapse (6 to 36 months)
8	Proto-colectomy (10 to 40 months)

TABLE 2.—*Results of Restoring Continuity (7 Patients)*

7	Re-united (24 to 36 months after by-pass)
3	Asymptomatic (6 months, 60 months, 96 months)
4	Recurrence (4 to 36 months)
2	Procto-colectomy
1	Re-by-passed
1	Remission on medical therapy

Four have relapsed with two undergoing total procto-colectomy, one patient has had re-diversion by ileostomy with remission of symptoms, and one patient is in remission on medical therapy. Although we have restored the colon successfully in only three of 24 patients to date, we remain enthusiastic about the initial response to diversion and have avoided permanent ileostomy in enough patients to justify further clinical trials of this form of therapy.

Sequelae of Operative Therapy

Aside from recurrent disease after operation, which we have already discussed, there are other problems or sequelae that may develop following operative therapy. These include both local and systemic problems.

Local problems

Ileostomy revisions may be required in patients undergoing total procto-colectomy. The problems encountered include ileostomy dysfunction due to prolapse, stenosis or herniation of a loop of adjacent ileum about the ileostomy. Recent technical improvements in the construction of ileostomy stomas has considerably reduced these problems. Local skin problems are unusual provided the patient receives expert exterostoma care.

Failure of the posterior wounds to heal after proctectomy has plagued most surgeons. This problem tends to occur more frequently in Crohn's disease. There is no universal agreement as to why these wounds fail to heal, although various technical errors have been postulated. Therapy for this complication has included the use of local antibiotics and silver nitrate application, re-excision of the sinus tract, removal of the coccyx, and the application of skin grafts. The prevention of these problems has been approached by primary closure of the perineal wound over suction catheters with some success and the endoanal approach that removes the sphincter but spares the levators with their fascia.

Systemic problems

Since many patients operated upon for ulcerative colitis and Crohn's disease have been receiving corticosteroid therapy for long periods, difficulty in discontinuing steroids is not uncommon. In such cases a gradual tapering of medication over a protracted period as well as the use of ACTH is sometimes necessary. Too rapid withdrawal is often followed by symptoms and signs of adrenal insufficiency—weakness, nausea, vomiting, fever, and postural hypertension.

Since vitamin B_{12} is absorbed in the distal ileum, when the terminal ileum is removed in Crohn's disease the patient should receive monthly vitamin B_{12} therapy to prevent macrocytic anemia.

Extensive small bowel resection may result in the "short bowel syndrome." Diarrhea or excessive ileostomy outputs leads to malabsorption and electrolyte losses. The use of Lomotil® or tincture of deodorized opium often slows intestinal transit. The diet should be augmented with vitamins and calcium and magnesium, if necessary. Excessive losses in these patients over long periods may lead to demineralization of the skeleton, formation of urinary calculi, anemia, and varying degrees of malnutrition.

Summary

The operation of choice in chronic *ulcerative colitis* is total procto-colectomy with permanent ileostomy. In selected patients total colectomy with ileorectal anastomosis may be indicated.

In patients with *regional enteritis* resection of the involved segment is preferred over by-pass by exclusion. "Skip lesions" proximal to the severely diseased segment may be left intact unless obstructing.

Diffuse Crohn's disease of the colon is best treated by total procto-colectomy with permanent ileostomy. Consideration should be given to diversion by ileostomy alone, particularly in young patients.

Segmental Crohn's disease of the right colon

may be treated successfully by local resection and ileo-transverse colostomy, whereas segmental disease of the transverse of left colon is best managed by total procto-colectomy.

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PART IV

The Ileostomy

Consideration of the Stoma, Peristomal Skin, Appliances and Diet

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IF MEDICAL MANAGEMENT of ulcerative colitis or regional enteritis has not been successful, surgical operation resulting in ileostomy may be necessary. Since toileting has been a personal, private matter since early childhood, the possible loss of control over this vital function is often overwhelming to the patient and his family. It is most important to provide the patient with a manageable ileostomy and teach him how to care for it. Stoma site selection, types of ileostomy, kinds of appliances, stoma and skin care, treatment of skin problems, and diet are important in the successful rehabilitation of the patient.

Once colectomy or by-pass ileostomy has been decided upon, choosing the stoma site is a consideration in the quality of the patient's life afterward. Various authors have described positioning of the stoma on the abdomen.[1,2] Certain criteria must be established before a decision can be made.

Since excrement may be liquid or soft and unformed and is eliminated day and night, an appliance must be worn 24 hours a day. It is therefore essential to place the stoma in an area manageable by the patient. He is the one who must care for it—often for life.

The site must be well away from all bony prominences—the costal margin, the superior anterior crest of the ilium and the symphysis pubis; and it must be such that the stoma will be visible to the patient when he is sitting or standing. Also it should be so chosen that articulation between the femur and the innominate bone will not interfere with wearing an appliance. Unusual contours, scarred areas and the umbilicus must be avoided. Although not usual in patients with ulcerative colitis or regional enteritis, obesity entails the additional restriction that the waist and abdominal folds also have to be avoided. Measurement for the stoma site should be made with the patient lying on his back, sitting up and standing, for a scar that is smooth with the patient supine may retract and pucker when he is sitting or standing. When the stoma site is not decided upon until the patient is in the operating room, this measurement in various positions is not done. Fortunately the iliac crest is most medial when the patient is supine, so the ileostomy usually is placed away from this most troublesome area. A well-healed pre-existing abdominal scar or the umbilicus causes far fewer problems than an ileostomy near the iliac crest.

Using these criteria, the stoma site usually is marked on the right side of the abdomen, below the waist, away from the iliac crest, the costal margin and the symphysis pubis and, in adults, at least two and a half inches from the umbilicus (see illustration). The ileostomy may be placed on the left side of the abdomen if the criteria can be better met there.

Stomas may be surgically matured or left nonmatured. The matured ileostomy is one that is constructed by bringing the ileum through the stoma site on the abdomen, trimming away the

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